



Case Report

Pontine cavernous haemangioma – An incidental autopsy finding

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ABSTRACT

Cavernous haemangiomas belong to a group of vascular malformations that are developmental defects of the vascular bed. Occurrence of an asymptomatic cavernoma in the pons is uncommon and worthy of record. At autopsy, cavernomas have to be differentiated from traumatic haemorrhagic lesions in head injury cases. We hereby report a case in which a pontine cavernous haemangioma was detected at autopsy in a 25-year-old female who died due to burn injuries.

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1. Introduction

Cavernous haemangiomas belong to a group of vascular malformations that are developmental defects of the vascular bed. These congenital abnormal vascular connections frequently enlarge over time. The lesions can occur on a familial basis. Patients may be asymptomatic, although they often present with headaches, seizures, or small parenchymal haemorrhages.¹ Pontine cavernous haemangiomas may be silent or manifest with gaze palsy, seizures or ataxia. Occurrence of an asymptomatic cavernoma in the pons is uncommon and worthy of record. We hereby report a case in which a pontine cavernous haemangioma was detected at autopsy in a 25-year-old female who died due to extensive burn injuries.

2. Case report

Medicolegal autopsy was conducted on the body of a 25-year-old female who died while on treatment for accidental burn injuries. The body weighed 48 kg and measured 156 cm in length. Infected burn injuries were present over the entire chest, abdomen, both arms and upper part of both thighs. The total body surface area burnt was estimated to be 65%. External examination did not reveal any other antemortem injuries on the body.

Internally, the lungs were oedematous with frothy exudate on cut section. The right and left pleural cavities and the peritoneal cavity contained approximately 400 ml, 360 ml and 330 ml of straw coloured fluid, respectively. The liver and both kidneys were congested. The uterus was unremarkable and was of parous type. Both ovaries contained multiple cysts which contained clear fluid. The brain was oedematous and weighed 1126 g. On sectioning the pons, a circumscribed reddish brown coloured lesion was observed towards the anterior portion measuring 1 cm in diameter (Fig. 1). Surrounding this, ferruginous penumbra was seen. Otherwise, the rest of the portion of brain stem and cerebellum was unremarkable.

Histopathology of the pons revealed a compact mass of dilated, thin walled blood vessels with absence of intervening brain tissue (Fig. 2A and B). Some of the vessels showed hyalinization of the wall. There was no direct arterial communication.

3. Discussion

Cavernous haemangiomas are an abnormal mass of thin walled vascular channels without intervening brain parenchyma.² They occur anywhere in the brain but are often supratentorial and involve the cerebral cortex. The incidence of these lesions is 0.4–0.5%.^{1,3} The cavernous haemangiomas in general show slight male preponderance and one fourth of the patients are children. Young adults most commonly present with seizures when cerebral cortex is affected. Haemorrhagic complications arise in about 1% of the cases. Brain stem haemangiomas may manifest with ataxia, diplopia, gaze palsy or may remain silent.⁴

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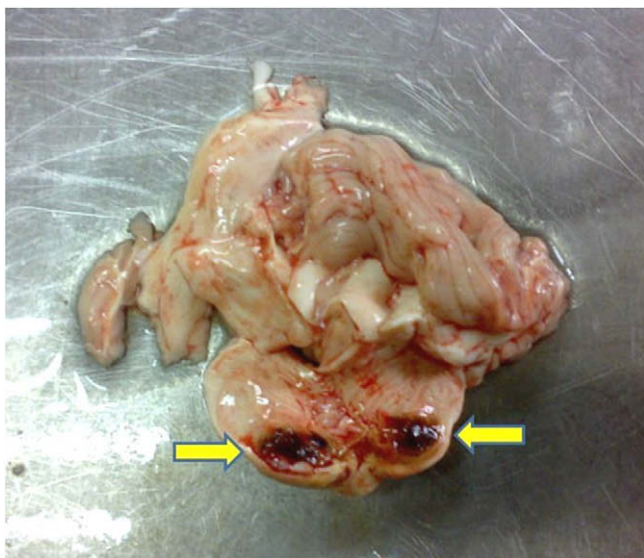


Fig. 1. Cut section of the pons showing cavernoma.

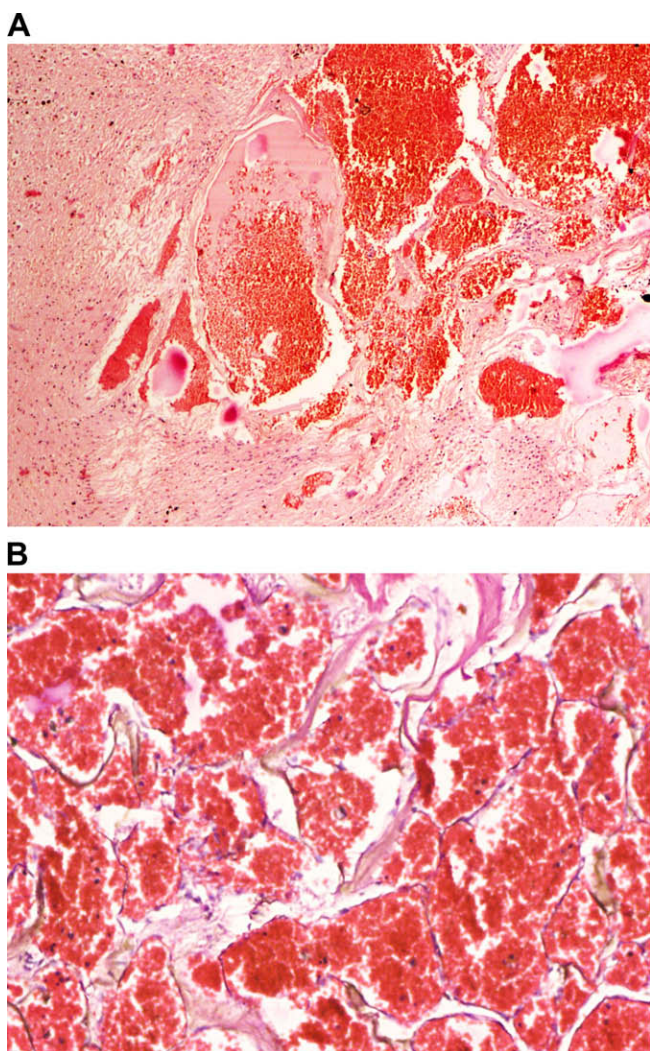


Fig. 2. (A) Section of pons showing pontine cavernoma and its relation with the brain matter, haematoxylin and eosin, original magnification 40 \times . (B) Section of pons showing dilated thin walled blood vessels without intervening brain tissue, haematoxylin and eosin, original magnification 100 \times .

Cavernous haemangiomas can be found in any part of the brain because they can occur at any location along the vascular bed.^{5–12} Frontal and temporal lobes are the most common sites of occurrence, and 80–90% of the lesions are supratentorial. The deep cerebral white matter, corticomedullary junction, and basal ganglia are common supratentorial sites, whereas the pons and cerebellar hemispheres are common posterior fossa sites. Intracranial extra cerebral cavernous haemangiomas also occur, but these are less common. They typically occur in the middle cranial fossa and originate from the cavernous sinus. Cavernous haemangiomas also can occur in the spinal cord, where they frequently coexist with multiple brain lesions. The differential diagnosis of these lesions in the brain is haemorrhagic tumors and resolving cerebral haemorrhage.

Although most cavernous haemangiomas are believed to be sporadic, many familial cases have been observed over the last two decades. These cases exhibit an autosomal dominant pattern of inheritance and seem to affect the Hispanic population in particular. Recent research has demonstrated at least three separate genes related to the familial form of the disease.¹³ The first gene is called *CCM1* (cerebral cavernous malformation 1) and is located on chromosome 7 at band 7q11.2–q21. The second gene is called *CCM2*. It is located at band 7p15–p13 and controls the production of a protein named malcavernin. The third gene (*CCM3*) identified as linked to familial cavernous haemangioma is on chromosome 3 at band 3q. Research is ongoing to further delineate the function of this gene and its relationship to cavernous haemangiomas. However, 40% can be linked to a *CCM1* genetic mutation and 20% can be linked to a *CCM2* mutation.²

With the advent of the MRI, the diagnosis of these lesions has increased. The surgical management of brain stem haemangiomas is controversial as focal neurological or neuro-ophthalmological complications are known to develop after surgery. Otherwise surgical excision is said to be curative, with residual neurological deficits improving over time. Conservative management with observation has also been reported to be successful. A newer option is the stereotactic radiosurgery, but this procedure is contraindicated in patients with concomitant venous haemangiomas.^{14,15}

Occurrence of an asymptomatic cavernous haemangioma of the pons is rare and worthy of record. Moreover, at medicolegal autopsy, cavernous haemangiomas have to be differentiated from traumatic haemorrhagic lesions in head injury cases. Such cavernomas may be grossly identified by their well circumscribed appearance with a reddish or bluish hue. However, histopathological confirmation remains the only way to identify these lesions at autopsy.

Conflict of Interest

The authors have no conflict of interest to declare.

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Ethical Approval

Not applicable.

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